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CORRESPONDENCE

Isolated cardiac sarcoidosis: Case experience in heart transplantation

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Sarcoidosis is a chronic immune disorder characterized by noncaseating granulomatous inflammation involving virtually every organ of the body and resulting in tissue destruction and impairment of organ function. More than 90% of patients have clinical presentations of asymptomatic hilar lymphadenopathy or pulmonary, cutaneous, or ocular involvement. Autopsy studies reported myocardial involvement in approximately 13–20% of the patients. The incidence of fatal cases was estimated at 25 per 100,000 but may be much higher in the Japanese population.[1] Isolated sarcoid heart disease without other systemic sarcoidosis precludes physicians from recognizing this subset of the population. The inflammation could be insidious, cause severe functional impairment of the heart or lethal arrhythmia, and even sudden death if the conduction system is affected. We present our experience with sarcoid heart disease mimicking idiopathic dilated cardiomyopathy in heart transplantation.

Three patients (one female, two males) age 29-47 years suffered from progressive dyspnea and leg edema with exercise intolerance. One patient had complete atrioventricular nodal and right bundle branch block shown on electrocardiogram. Echocardiogram showed four-chamber dilatation and global hypokinesia with regional wall abnormalities. Computed tomography of the chest in one patient confirmed no evidence of bilateral hilar lymphadenopathy. Idiopathic dilated cardiomyopathy was diagnosed in all three patients; they underwent orthotopic heart transplantation. The explanted hearts consisted of generally dilated ventricles with patent coronary arteries (Fig. 1A). Extensive fibrosis of the left ventricle and the interventricular septum was seen in all patients. Microscopically, pancardiac involvement by noncaseating granulomatous inflammation (Fig. 1B) was predominant in the left ventricle and interventricular septum. Sections showed scattered multinuclear giant cells with occasional eosinophils, mimicking the clinical picture of giant cell myocarditis (Fig. 1C). Cytoplasmic asteroid bodies can be discernible in giant cells (Fig. 1C, inset), which also encased large vessels and induced vasculitis (Fig. 1D). Immunohistochemically, the giant cells stained for CD68 antibody but were negative for smooth-muscle actin. The posttransplant course was all uneventful.

Cardiac sarcoidosis is often overlooked clinically, particularly if the effect on the heart is predominant or isolated. In 1977, Roberts et al[2] described that most patients in their necropsy study who died from cardiac sarcoidosis lacked clinical features of extracardiac

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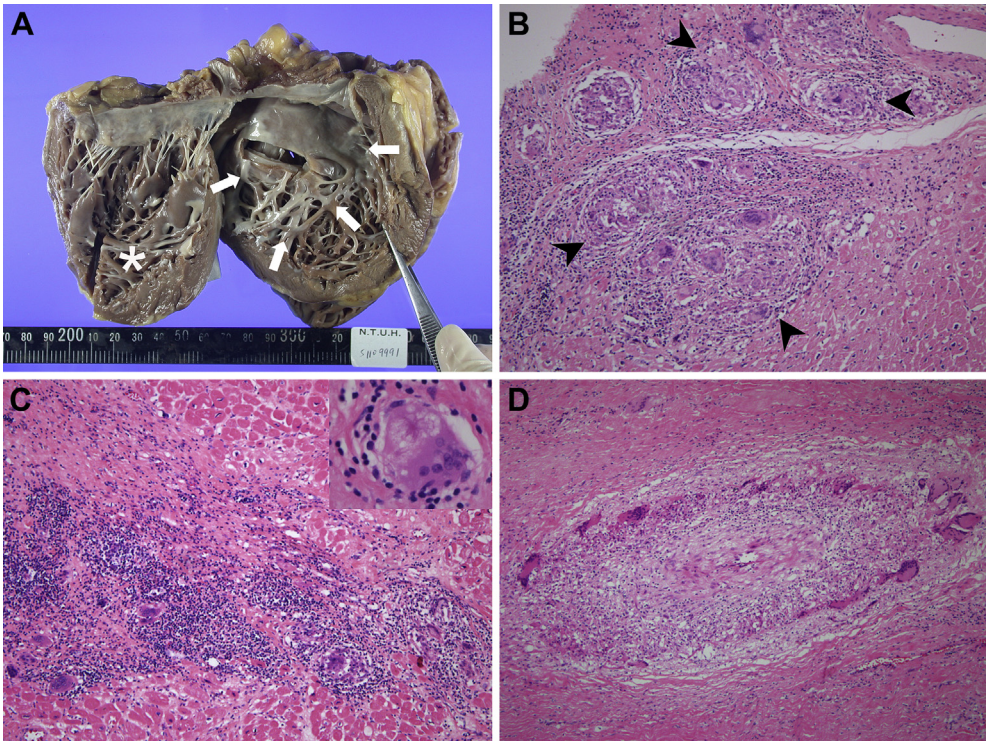

Figure 1 (A) Gross specimen of right ventricle from case 3. There was extensive fibrosis of the interventricular septum (arrows), with the asterisk indicating relative sparing of the right ventricle. (B) The arrowheads point to noncaseating granulomatous inflammation with destruction of myocardium (hematoxylin and eosin stain, ×100). (C) High-power magnification shows scattered multinuclear giant cells between necrotic myofibers resembling the clinical picture of giant cell myocarditis, which may be misidentified with endocardial biopsy (hematoxylin and eosin stain, ×200). Many cytoplasmic asteroid bodies are found in all three cases (inset, hematoxylin and eosin stain, ×400). (D) Vasculitis induced by multinucleated giant cells with destruction of vascular wall (hematoxylin and eosin stain, ×100).

disease. Because of left side predominance, cardiac sarcoidosis is often difficult to diagnose with right ventricle endomyocardial biopsy. Therefore, the guidelines published by the Japanese Society of Sarcoidosis and Other Granulomatous Disorders in 2006 suggest clinical cardiac involvement by sarcoidosis can be otherwise established by demonstration of clinical signs and symptoms and/or abnormalities seen on electrocardiogram, echocardiogram, and other imaging studies associated with extracardiac sarcoidosis evidence.[3]

Cardiac sarcoidosis is a potentially controllable disease with steroid and antiarrhythmic devices that deserves wider recognition for its occurrence. With early diagnosis, deterioration of cardiac sarcoidosis might be prevented by corticosteroid therapy.[4,5] Heart transplantation offers a rescue therapy for severe heart failure or refractory arrhythmia, and the prognosis of transplantation in cardiac sarcoidosis seems to be favorable in 1-year survival but has not been completely verified. Recurrence of granulomas in the allografts was reported, and these are treated with long-term steroid and immunosuppressant therapy.

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